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The diagnosis of dementia in persons with mental retardation: validating methods of assessment

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THE DIAGNOSIS OF DEMENTIA IN PERSONS WITH MENTAL RETARDATION: VALIDATING METHODS OF ASSESSMENT

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ABSTRACT

The usual assessment instruments for the diagnosis of dementia are often difficult to use when evaluating the disorder among individuals with mental retardation. This study investigates whether a modified method based on Visser et al. (1997) can identify a dementing process. Ninety individuals diagnosed with severe and profound mental retardation were studied. One half of the participants were diagnosed with Down's syndrome. The participants were equally assigned to one of three groups based on perceived risk for dementia. A differential prevalence design was used. Both cross sectional and longitudinal analyses were utilized in this study. Results indicate that the Visser et al. (1997) method is effective in identifying dementia. Differences between syndrome and risk of dementia groups will be discussed.

Introduction

A significant growth in the elderly population with mental retardation (MR) has been noted in recent years (Deb & Janicki, 1995; Janicki, 1994; World Health Organization, 2000). With this increase, there is a greater proportion of older persons who develop age related changes affecting their cognitive, physical, emotional and adaptive functioning (Cherry, Matson & Paclawsky, 1997; Kapell et al., 1998). These physical differences include failing senses, lessened attentiveness, declining agility, difficulties with mobility, reduced resistance to disease, difficulty in recovering from illness and injury, decreased stamina, depression and anxiety (Anderson, 1993; Evenhuis et al., 2001; Moss et al., 1993; Williams, 1995). Increases in the prevalence of age associated psychiatric diseases such as Alzheimer's disease and other dementias have also been noted. Researchers have shown that individuals with mental retardation, particularly those diagnosed with Down's syndrome, are at an even greater risk for dementia than the general population (Burns, 1992; Zigman, Silverman & Wisniewski, 1996).

Several types of dementia have been identified (i.e., Alzheimer's disease, vascular dementia, frontal lobe dementia, subcortical dementia and dementia due to HIV or

other general medical conditions). Some forms of the disorder are reversible (i.e., dementia caused by hypothyroidism, drug induced dementia)(Coffey & Cummings, 1994), therefore careful assessment is important to distinguish irreversible dementia from reversible dementia or dementia due to a treatable condition (Kaplan, Sadock, & Grebb, 1994). Best practices for the assessment of dementia among individuals in the general population using clinical interview, physical exams, and neurological testing are clearly established and have been conducted for many years (Coffey & Cummings, 1994). However, the standard of care in diagnosis of dementia among individuals with mental retardation is less clear. The diagnosis of dementia in persons with developmental disabilities, especially in the early stages, is made difficult by the lack of reliable and standardized criteria and diagnostic procedures and the trouble of detecting declines against the already impaired background of developmental disability (Aylward, Burt, & Thorpe, 1997). Thus far, the DSM-IV (APA, 1994) does not provide any information regarding how to diagnose aging individuals with dementia and mental retardation. Perhaps this state of affairs can be attributed to standard methods of dementia assessment (i.e., cognitive evaluations, mental status examinations,

and neuropsychological tests) that appear to be inappropriate to diagnose persons who have presented with cognitive delays since childhood (Haveman, Maaskant & von Schrojenstein, 1994). Therefore, researchers have begun to address the issue of diagnosing dementia in individuals with developmental delays.

Aging in the Developmentally Disabled

The number of individuals that comprise the "elderly population" is rapidly on the rise (Janicki, Dalton, Henderson & Davidson, 1999). According to the 2000 US Census, there are approximately 35 million people age 65 years or older. This dramatic increase in life expectancy can be attributed to advances in medicine, public health, science, education and technology (World Health Organization, 2000).

Additionally, owing to more readily available supportive services to individuals with mental retardation (MR), a much greater proportion of persons with developmental disabilities are reaching old age (Bittles et al., 2002; Das & Mishra, 1995). For instance, persons with Down's syndrome had a life expectancy of less than 10 years in 1929, but with advances in cardiac care and surgical procedures as well as the availability of general community health care systems, the life expectancies of these

individuals have significantly increased (Zigman, Schupf, Haverman & Silverman, 1995). Over 40% of live-born Down's infants now survive 60 years (Holland, 1999; Yang, Rasmussen & Friedman, 2002). The life expectancies of non-Down's syndrome persons with MR have dramatically changed too. The average age of death for persons with intellectual disabilities in 1931 was 22 years. This age had increased to 59 years by 1976 and to 66.1 years by 1993 (Janicki et al., 1999). Researchers estimate that, worldwide, there are as many as 60 million persons who currently have some level of developmentally related cognitive impairment (World Health Organization, 2000) and there are between 200,000 and 500,000 older adults with MR in the United States (Janicki & Dalton, 2000). This older age group will likely double by the year 2025 (Rehabilitation Research and Training Center, 1995). In fact, some researchers believe that this "old age" MR population could reach anywhere between 700,000 to 4,000,000 in the next 20 to 30 years (Silverman et al, 1998). As these survival rates are gradually approaching that of the general population, researchers have begun to focus on age-related health conditions (e.g., physical and functional changes) among persons with MR.

Physiological Changes

Researchers have found that the combination of mental retardation and aging may create additional difficulties for an individual (LeBlanc & Matson, 1997). Campbell and Herge (2000) advocate that individuals with MR often experience a need for additional support due to physiological and psychological changes associated with aging. Common age related physical changes that are experienced by both the general and developmentally disabled populations include hearing and visual impairment, decreased muscle mass and flexibility, and increased incidence of arthritis, hypertension, heart disease, diabetes, and osteoporosis (Anderson, 1993; Evenhuis et al., 2001; Janicki & Dalton, 1998; Moss et al., 1993). Although these conditions are common to both the developmentally disabled and the general populations, individuals with mental retardation have a higher incidence of death, disease, and disability. For instance, persons with mental retardation have been identified as a "population at risk" because of their poor nutrition decisions and sedentary lifestyle (Petitti & Campbell, 1991). Although researchers have found that a third of all Americans are overweight, obesity among individuals with mental retardation (particularly females) is even higher

(US Department of Health and Human Services, 2002). In fact, close to half of all people with developmental delays are overweight (Special Olympic Report, 2001). The prevalence of obesity increases among individuals with Down's syndrome. Prasher and Chung (1996) found that 52% of their participants who were diagnosed with Down's syndrome were clinically obese. Braunschweig and colleagues (2004) reported that 89% of Down's syndrome participants in their study on nutrition were overweight or obese. The high levels of excess fat found in people with developmental disabilities expose them to a higher risk for many different types of disease associated with obesity. Researchers have also noted that individuals with mental retardation are at risk for various diseases (e.g., vascular disease, diabetes, hypertensive encephalopathy, etc.) that result from physical inactivity (Rimmer, 1994).

Researchers have noted that some underlying characteristics and commonalities typical to individuals with mental retardation may also have adverse effects on their health. For example, individuals with cognitive disabilities may have difficulty accessing emergency health services (Spreat & Conroy, 2001). Persons with communication deficits are likely to have difficulty communicating with health care providers (Moss, 1995).

Also, researchers have found that people with mental retardation are more likely to be prescribed medication than individuals in the general population (Kumar & Brecher, 1999; Young & Hawkins, 2002). If these individuals, rather than their caretakers, are responsible for following their medication regime, rates of medication non-compliance due to forgetfulness increases drastically (Torr & Chiu, 2002). Also, people with a long life history of taking certain medications (e.g., anticonvulsants, neuroleptics) are at a higher risk of developing secondary conditions (e.g., osteoporosis, tardive dyskinesia) (Zubenko & Sunderland, 2000). Finally, several disabilities have been found to be associated with their primary diagnosis and syndromes (e.g., Down's syndrome is associated with a high incidence of thyroid disorder, cardiac myopathy, and senile dementia of the Alzheimer's type).

Psychological Changes

Similarly to physical health, there are also psychological issues related to aging. Historically, many professionals believed that persons with mental retardation were incapable of developing emotional problems because of their lack of "proper ego strength" (Reiss, Levitan, & Szyszko, 1982). Practitioners assumed that odd or strange

behaviors in persons with developmental disabilities were simply due to the individual's cognitive limitations. The term "diagnostic overshadowing" was coined in 1982 because mental retardation was said to "overshadow" the symptoms of psychological disturbance (Reiss, 1994; Reiss, Levitan, & Szyszko, 1982).

Experts in the field have made great strides in the past two decades in identifying and diagnosing mental illness in persons with mental retardation. As a result, it has been well documented that individuals with developmental delays are susceptible to the full range of emotional and personality disorders (Davidson et al., 1994; Matson & Barrett, 1993; Reiss, 1994). Additionally, the data gathered suggests that the prevalence of these disorders in the mentally retarded population is higher than that of the general population (Dudley, Ahlgrim-Delzel & Calhoun, 1999; Matson & Sevin, 1994; Moss et al., 1997; Rojahn & Tasse, 1996). Prevalence rates of psychopathology ranging from 10 to 60% percent have been reported (Borthwick-Duffy, 1994; Davidson et al., 1994; Jacobson, 1999; Matson & Barrett, 1994). Despite the awareness of high rates of co-morbid psychiatric conditions among individuals with MR, researchers continue to show that behavioral and emotional difficulties are less likely to be

accurately acknowledged compared to disorders of persons of average intelligence (Jopp & Keys, 2001). White et al. (1995) estimated that people with MR could expect a 19 percent drop in diagnostic accuracy and mental health treatment recommendations in contrast to persons with comparable symptoms who do not have other disabilities.

Although attempts to accurately diagnose individuals with developmental disabilities and co-morbid psychiatric problems continue to be substandard, researchers have demonstrated that the rate of psychopathology among persons with MR remains stable throughout their life span. Cherry, Matson, and Packlawskyj (1997) investigated the prevalence of psychopathology in individuals with severe and profound mental retardation and found that the frequency of disorders was comparable for younger and older adults but older adults showed longer duration and/or greater severity ratings than did younger adults. The mental health problems prevalent in older individuals with mental retardation are consistent with those commonly found among aging persons of normal intelligence (i.e., anxiety, phobias, and depression) (Harper & Wadsworth, 1990; McNellis, 1997; Rojahn, Warren, & Ohringer, 1994). Researchers have hypothesized that these psychiatric conditions can result from physiological changes, medical conditions, long term

pharmaceutical use, or changes in living situation or lifestyle (Moss et al., 1997). In addition to high rates of mood, anxiety, and adjustment disorders, researchers have noted an increase in the prevalence of age-associated psychiatric disorders among persons with mental retardation. One such condition is dementia associated with old age.

Dementia

Dementia is a syndrome where progressive deterioration in cognitive abilities is sufficiently severe to interfere with the individual's usual social or occupational functioning or their level of personal adjustment (APA, 1994). This condition is neurologically based and requires the presence of memory problems plus one of the following: aphasia (difficulty in expressing thoughts as spoken words), apraxia (difficulty in carrying out simple, directed acts), agnosia (difficulty in interpreting familiar faces or other well-known objects), and disturbances in executive functioning (the ability to plan and organize). Eventually, this problem has dramatic effects on how well a person is able to care for him/herself. An estimated 5-15% of people 65 years of age and older are affected by a dementing disorder (Kaplan, Sadock, & Grebb, 1991; Riley, 1999). Researchers have

found that several risk factors are associated with dementia. These variables include a family history of dementia, low educational level, previous head trauma, cardiovascular disease, stroke, diabetes, apolipoprotein E-4 allele and previous major depressive episode (Ravaglia, 2002; Sliwinski, Buschke, Stewert, & Masur, 1997).

Most adults with mental retardation are at the same risk for dementia as are older adults in the general population (McNellis, 1997; Zigman et al., 2004). Thus far, the only particular risk factor uniquely identified among people with mental retardation is Down's syndrome (Dalton & Janicki, 1999). Researchers have found that people with Down's syndrome have significantly higher rates of dementia of the Alzheimer's type (Holland, Hon, Hubert & Stevens, 2000; Zigman, Silverman & Wisniewski, 1996) and prevalence increases in an exponential fashion past age 50. Approximately 25% of adults with Down's syndrome age 40 to 49 years, approximately 55% of those age 50 to 59 and about 75% of adults age 60 years and older show the behavioral symptoms of dementia (Zigman, Schupf & Haveman, 1997).

Although not every aging individual with Down's syndrome goes on to show clinical manifestations of dementia (Devenney, Silverman, Hill, Jenkins, Sersen, & Wisniewski, 1996), upon autopsy, virtually all adults with

Down's syndrome over the age of 40 show some evidence of the neuropathology associated with Alzheimer's disease (Mann, 1993). For instance, these individuals demonstrate deposition of beta-amyloid in diffuse and neuritic plaques and intracellular neurofibrillary tangles (Royston et al., 1999). This phenomenon appears to be due to the triplication of the genes for beta-amyloid precursor protein (B-APP), located on the proximal part of the long arm of chromosome 21 (Rumble et al., 1989; Schupf, 2002). Thus far, it is not clear what effect, if any, possible risk factors as seen in the general population have on people with Down's syndrome (Zigman, Schupf & Haveman, 1997).

Although Alzheimer's disease is the most common and most researched form of dementia, dementing disorders are a heterogeneous group of conditions. Their etiology, neurological substrate, disease course, and treatment can vary greatly (Coffey & Cummings, 1994). Twelve categories of dementia are outlined in DSM-IV. They are Dementia of the Alzheimer's type, Vascular Dementia, Dementia due to HIV Disease, Dementia due to Head Trauma, Dementia due to Parkinson's Disease, Dementia due to Huntington's Disease, Dementia due to Pick's Disease, Dementia due to Creuitzfeldt-Jakob Disease, Substance Induced Persisting

Dementia, Dementia due to Multiple Etiologies, and Dementia due to Other Medical Condition (APA, 1994). Several types of dementia will be discussed next.

Alzheimer's Disease

Alzheimer's disease is the most common form of dementia – an illness associated with old age that impairs intellectual functions (e.g., memory, orientation, concentration, language, perception, executive functioning)(Schofield & Mayeux, 1998). This disorder is a slow and progressive, degenerative disorder of the brain that eventually results in diminished brain function and death. According to prevalence data, approximately 100,000 victims die and 360,000 new cases are diagnosed each year (Small et al., 1997). Researchers estimate that by 2050, 14 million Americans will have this disease (Cummings & Cole, 2002). As mentioned, Alzheimer's disease is an age-associated disorder but it is not a part of normal aging. The disease usually begins later in life (late onset – after age 60); however, it may affect persons as young as 30 years of age (early onset). Early onset AD results in about 5-10% of all cases (Cummings & Cole, 2002) and typically has a much faster course and progression of deterioration.

In the early stages of AD, the symptoms may be subtle. New or recent memory is typically impaired first, and the individual may find it hard to learn and retain new information. Eventually, older or distant memory also is lost. Next, other symptoms may appear, including aphasia, apraxia, agnosia, and disturbances in executive functioning (Grabowski & Damasio, 1997). In practical terms, individuals with early AD commonly misplace things, frequently repeat statements, have trouble finding names for familiar objects, get lost on familiar routes, and lose interest in things they previously enjoyed. However, despite all of these intellectual problems, Lowenstein (1990) found that many people with early Alzheimer's disease continue to be able to eat, bathe, dress and groom themselves as usual without assistance.

Unfortunately, psychiatric symptoms (personality changes, irritability, anxiety, and depression) also may occur, and these may cause serious problems in relationships with family and friends (Balestreri, Grossberg, & Grossberg, 2000; Hargrave, Stoeklin, Haan, & Reed, 2000). Personality changes have not been well specified and thus, have often gone unrecognized. However, in recent years, it has become more widely known that some dementias can cause changes to an individual’s core

personality and social-affective functioning. Researchers have demonstrated that personality change is a significant predictor of dementia, independent of cognition and functional status. Smith-Gamble et al. (2002) assessed the predictive value of caregiver reports of changes in personality on incident dementia and Alzheimer’s disease. Subjects with changes in personality had approximately twice the odds of having dementia as subjects with no change in personality.

As Alzheimer's disease progresses to its middle and late stages, there may be delusions (irrational beliefs, especially about being persecuted or having one's belongings stolen) and hallucinations (false sensations — seeing, hearing, smelling, tasting or being touched by something that isn't really there). The patient also may become aggressive or may begin to wander away from home if left alone (Riley, 1999). In the final stages of AD, a person can no longer function without assistance. Most people in this stage no longer understand language, they no longer recognize family members, and they can no longer perform basic activities of daily living such as eating, dressing, and bathing (Ballard et al., 2001).

The rate that AD advances is different for each person. If AD develops rapidly, it is likely to continue

to progress rapidly. If the disease has been slow to progress, it will likely continue on a slow course. The duration of the illness may often vary from 3 to 20 years. The assessment of dementia will soon be discussed but it is important to note that the diagnosis of Alzheimer's is always a diagnosis of exclusion. It is made based on characteristic symptoms and by excluding the other causes of dementia. The diagnosis of Alzheimer's disease can only be confirmed by microscopic examination of a sample of brain tissue after death (Rogan & Lippa, 2002).

Vascular Dementia

Vascular dementia (VD) is the second most common cause of dementia, accounting for approximately 20% of all cases alone and up to another 20% of cases in combination with Alzheimer's disease (Pantoni & Inzitari, 2002). VD is not a single disease but a group of syndromes associated with problems in circulation of blood to the brain (cerebra-vascular disease). Some subtypes of VD include: (1) multi-infarct dementia, (2) VD due to strategic single infarct, (3) VD due to lacunar lesions, (4) VD due to hemorrhagic lesions, and (5) Binswanger disease.

VD is characterized by uneven impairment in cognitive functioning and "patchy" performance on neuropsychological measures (Jefferson et al., 2002). For instance,

individuals often demonstrate preserved ability on some domains but impaired performance on others depending on the site of the infarct or infarcts (Coffey & Cummings, 1994). Unlike Alzheimer's disease, the onset of cognitive impairment is often abrupt and a "stepwise" decline in cognitive functioning is typically observed as the disease progresses, with each step representing the occurrence of another vascular event. VD usually affects people between the ages of 60 and 75 years and is slightly more common in men than women (Alexopoulos, 2003). Individuals who have had a stroke are at increased risk for vascular dementia. In fact, researchers have demonstrated that the prevalence of dementia is nine times greater in patients who have had a stroke than in controls. One year after a stroke, approximately 25% of patients develop new onset dementia. Within four years following a stroke, the relative risk of incident dementia is 5.5 times more likely for these individuals than for persons who do not have a history of stroke (Madureira, Guerreiro, & Ferro, 2001).

Frontal Lobe Dementia

Frontal Lobe Dementia (FLD) is the name given to any dementia caused by damage to the frontal lobe portion of the brain. This part of the brain is known to govern mood, behavior, judgment, and self-control. FLD includes Pick's

disease, but can be caused by other disorders. Like Alzheimer's disease, Pick's disease dementia causes a progressive and irreversible decline in a person's abilities (Litvan, 2001). From the onset of the disease, life expectancy is 2 to 15 years, with an average of 6 to 12 years. The first symptoms are typically psychological and behavioral problems. In fact, the diagnosis is often suspected to be a psychiatric illness. Individuals with FLD demonstrate deterioration of social skills and changes in personality early in the course of the illness yet they lack insight into the effects of their behaviors (e.g., show insensitivity to others, emotional blunting, behavioral disinhibition) (Kertesz, 2000). The individual often becomes "obsessional" in these early stages, repeatedly washing hands, observing little rituals, or insisting that everything is in order (Worthington, 1996). FLD is also characterized by prominent language abnormalities. For instance, the individual may begin using pat phrases repeatedly and excessively, lack spontaneous speech, and demonstrate a decrease in vocabulary. Eventually, the individual's dialogue is unintelligible and they may become completely mute by the end of the disease (Hodges, 2001).

Pick's Disease can affect both men and women and it typically begins affecting people between 40 and 65 years of age (Riley, 1999). As with Alzheimer's disease, the cause cannot be determined in most cases; however there are strong genetic components in certain families. A mutation on chromosome 17 has been identified and this genetic component has been described as affecting 20 to 50% of people with Pick's disease (Bird, 1998).

Subcortical Dementias

The subcortical dementias are a group of disorders characterized by primary dysfunction in the subcortical areas of the brain. These dementing conditions lead to motor dysfunction, speech impairment, memory dysfunction, executive disorders, and disturbances in mood and personality (Markesberry, ed., 1998). Subcortical dementia occurs with extrapyramidal syndromes such as Parkinson's disease, Wilson's disease, progressive supranuclear palsy, and Huntington's disease. Although there is often a general decline in intellectual processes over time, this decline is usually much less severe than in other dementing disorders. Parkinson's and Huntington's diseases will be briefly described.

Parkinson's Disease. Parkinson's disease is a slow, progressive neurological condition characterized by tremor,

rigidity, bradykinesia, and postural instability. Dementia is noted in 20 to 60% of these cases (Aarsland et al., 2001). Cognitive and motoric slowing, executive dysfunction, and impairment in memory retrieval that is often exacerbated by depression characterize dementia associated with this disease (Ebmeier, Calder, Crawford, & Stewart, 1990).

Huntington's Disease. Huntington's disease is an inherited, progressive, degenerative disease of cognition, emotion, and movement. This condition is usually diagnosed in the late 30's or early 40's but may begin as early as age 4 or as late as age 85. Onset is often noted to have changes in behavior and personality (inclusive of depression, irritability, and anxiety). Some individuals present with abnormalities of movement that resemble increased fidgeting and later progress to the characteristic "generalized choreothatosis". Difficulty with memory retrieval, executive functioning, and judgment are common (Paulsen et al., 2001). Disorganized speech and psychotic features are sometimes present. Late in the disease, marked ventricular dilation consistent with advanced cerebral atrophy (a.k.a., "boxcar ventricles") may be seen on structural brain imaging.

Clinical Evaluation of Dementia

For a diagnosis of dementia, current criteria in the Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV), require evidence of decline from previous levels of functioning and impairment in multiple cognitive domains, not solely memory (APA, 1994). In order to do this, clinicians have used a combination of clinical interviews, medical evaluations, and neuropsychology measures to detect cognitive and memory changes within the general population. These methods will be discussed next.

Clinical Interview

A person's medical and psychosocial history is an important part of the dementia evaluation. Taking a thorough history involves gathering information from the individual, as well as the person's family members and friends. Careful questioning is required to elicit clues to the presence of functional and cognitive impairment (Zelinski, Gilewski & Schaie, 1993). Questions should be asked about forgetfulness and orientation. Information about the individual's daily functioning should also be obtained. For example, it is important to ask questions about the individual's ability to do activities like take care of their hygiene, prepare meals, pay bills, remember appointments, take medications, and travel out of the

neighborhood (APA Presidential Task Force, 1997). An informant can be very helpful in providing information about the person's symptoms, such as when the symptoms were first noticed, how quickly they developed, and whether they have continued to get worse. The interview must review the individual's past medical, social, educational, and vocational histories; whether there is a history of dementia in the family; and whether there have been any recent unusual events in the person's life. Any significant exposure to alcohol, medications, and other possible toxins must also be considered (Brooke, 2002).

Physical Exam and Laboratory Results

Current DSM-IV criteria require evidence of impairment that interferes with the person's previous level of social and occupational functioning. However, because medications and various medical disorders may have adverse effects on an individual's baseline functioning, a thorough assessment for chronic disease processes and a medication review is necessary to rule out specific treatable causes of dementia. Disorders found to cause symptoms of dementia include hearing or vision deficits, hypothyroidism, vitamin B_{12} deficiency and depression (Rogan & Lippa, 2002). These disorders are relatively easy to detect therefore

appropriate laboratory tests, physical examinations, and psychological tests should be administered.

Laboratory tests recommended for the diagnostic work-up of dementia include a complete blood cell count (to exclude anemia and infection) and urinalysis (to exclude infection). Serum electrolyte, glucose and calcium levels, blood urea nitrogen, serum creatinine level and liver function tests should also be done to investigate metabolic disease (Quality Standards Subcommittee, 1994). Syphilis serology, erythrocyte sedimentation rate, serum folate level, human immunodeficiency virus (HIV) status, urine check for heavy metals and toxicology screening may be indicated in a minority of cases (Rabins, Lyketsos & Steele, 1999). Lumbar puncture is usually not necessary except when the onset of dementia occurs before 55 years of age or when a specific condition such as infection, syphilis or vasculitis is suspected (Quality Standards Subcommittee, 1994). The physical examination should also include assessment of cognitive domains, including speech (aphasia), motor memory (apraxia), sensory recognition (agnosia) and complex behavior sequencing (executive functioning) (Kramer & Duffy, 1996). This often requires referral for neuropsychological assessment (Drane & Osato, 1997).

Aphasia is a disorder that results from damage to language centers of the brain. It can also occur with damage to the connections between centers of transcortical aphasia. Some individuals with aphasia have difficulty with expressive language (what is said) while others have problems with receptive language (what is understood). Language can be affected not only in its oral form (talking and comprehending) but also in its written form (reading and writing). Word finding problems (dysnomia/anomia) are common in people with aphasia. Asking the individual to name body parts or objects in the room may informally assess aphasia. Frequent use of vague terms such as "thing" and "it" may also signify deterioration of language function (Kertesz, 1994).

Apraxia is a motor disorder of action planning in which volitional or voluntary movement is impaired without muscle weakness. Heilman, Watson and Rothi (2000) defined apraxia as a disorder of skilled movement not caused by weakness, akinesia, deafferentation, abnormal tone or posture, movement disorders such as tremors or chorea, poor comprehension, or uncooperativeness. An example of a test for apraxia is to ask the patient to pantomime the use of a common object such as a hammer or a toothbrush (Taylor, 1994).

Agnosia is defined as the loss of ability to perceive or recognize sensory stimuli (i.e., objects, people, sounds, shapes or smells). One way to assess for agnosia is by first asking the patient to close his or her eyes and then placing an object, such as a key or a coin, in the patient's hand and asking the patient to identify it without looking at it. Ringing a bell or honking a horn can evaluate auditory agnosia. Asking the individual to identify a common scent (i.e., vanilla or lemon) can test olfactory agnosia. Inability to recognize a common object despite normal sensory thresholds signifies agnosia (Kramer & Duffy, 1996).

Osborn (1998) defined executive functioning as the ability to organize thought and work, to create plans and successfully execute them, and to manage the administrative functions of one’s life. Asking the patient to perform a series of simple tasks is a way to evaluate executive functioning. For example, the individual can be asked to sign a piece of paper, put the piece of paper in his or her right hand, fold it in half and put it on the floor. This task would be difficult for a person with impairment in the ability to plan, initiate, sequence and monitor complex behavior. Asking the person to perform serial subtraction of 7s (backward from 100 to 65), to spell the word "world"

backward and to produce verbal word lists, such as names of animals or items in a grocery store, are other ways to test executive functioning and abstract thinking.

Neuropsychological Tests

Neuropsychological tests are often administered to assess difficulties in attention span, perception, memory, problem solving, and social and language skills. These tests are often used to screen for cognitive impairment that may be indicative of a dementing process. An individual's responses on a neuropsychological battery may also provide clues to the underlying cause of dementia. For example, Jefferson et al. (2002) found that individuals with Alzheimer's disease showed differential impairment on the Mini Mental State Examination (MMSE) indices measuring orientation and memory when compared to individuals with ischemic vascular dementia (IVD) and Parkinson's disease (PD). The IVD and PD groups performed significantly worse than the persons with AD on the MMSE indices assessing working memory and motor/constructional functions. The authors therefore concluded that these indices could assist clinicians in deriving important information regarding the etiology of a patient's dementing illness.

Common neuropsychological tests used for the assessment of dementia include: Folstein's Mini Mental

State Examination (1975), the Neurobehavioral Cognitive Status Examination (1989), and the Mattis Dementia Rating Scale (1976, 1988).

The Mini Mental State Examination (MMSE; Folstein, 1976) is a brief, quantitative measure that has been widely used for assessing cognitive mental status. This test can be used to screen for cognitive impairment, to estimate the severity of mental impairment at any given point in time, to follow the course of cognitive changes in an adult over time, and to document an individual's response to treatment. The MMSE has demonstrated validity and reliability in psychiatric, geriatric, neurologic, and other medical populations (Mitrushina & Saltz, 1991; Tombaugh et al., 1996). However, studies have shown that it has limited specificity with respect to individual clinical syndromes (e.g., dementia or delirium) (Kirby et al., 2001; Tierney et al., 1997; Sabe, Jason, Juejati, & Leiguarda, 1993). The test assesses orientation, attention, immediate and short-term recall, language, and the ability to follow simple verbal and written commands. Furthermore, it provides a total score that places the individual on a scale of cognitive function. The maximum MMSE score is 30 points, individuals with a score of 23 may

be experiencing dementia (Sabe, Jason, Juejati, & Leiguarda, 1993).

The Neurobehavioral Cognitive Status Examination (CSE; 1989) is a standardized test instrument for assessing cognition. It is designed to rapidly assess intellectual functioning in five major areas: language, constructional ability, memory, calculation skills, and reasoning/judgment. Three general factors are also examined: level of consciousness, attention, and orientation. The test requires less than 5 minutes to administer to individuals with “normal” functioning, and 10-20 minutes for those who are impaired. The NCSE can be compared to the Mini-Mental State Exam (MMSE). Studies investigating the reliability of the NCSE demonstrate good test-retest reliability, but poor inter-rater reliability (Lamarre & Patten, 1994). Few independent validation studies are available for this instrument. Drane and Osato (1997) found the NCSE can be used to successfully identify cognitive dysfunction in all patients with a diagnosis of dementia (high sensitivity) but there were major problems with the specificity of the test. The researchers found the NCSE to generate an unacceptable level of false positives among the healthy older adults (70%). Therefore,

this test should not be used alone as a screening instrument for dementia.

The Dementia Rating Scale (DRS; Mattis, 1976, 1988) is a well-known and widely used instrument for assessing dementia. It consists of five subscales that evaluate attention, perseveration and initiation, construction, memory, and conceptualization. The DRS is composed of simpler items than traditional cognitive tests, thus decreasing its susceptibility to floor effects and increasing its sensitivity to individuals with substantial cognitive defects (Marson, Dymek, Duke, & Harrell, 1997). It is brief and easy to administer, typically taking between 20 to 40 minutes. Researchers have demonstrated that that the scale has excellent test-retest and inter-consistency reliability (Vitaliano et al., 1984) as well as construct and criterion related validity (Vitaliano et al., 1984; Vitaliano, Russo, & Breen, 1986). The DRS total score also appears to validly quantify cognitive impairment for individuals with dementia (Moss & Alberts, 1988; Shay et al., 1991). The DRS also can be used to track changes in cognitive status over time.

The Assessment of Dementia among Persons with MR

The standard clinical criteria for diagnosing dementia may be inadequate for assessing individuals with

developmental disabilities. Often, individuals with MR do not present the traditional clinical signs of dementia (i.e., cognitive deterioration) or these signs may be difficult to detect (Menolascino & Potter, 1989). For instance, individuals with developmental disabilities often demonstrate cognitive deficits such as memory impairment, receptive/expressive communication delays, and executive functioning disturbances at an early age. The mere presence of these cognitive deficits does not equate to a diagnosis of dementia because these impairments may have been present throughout the persons' life (Haveman et al., 1994).

Therefore, in persons with mental retardation, a diagnosis of dementia should be made based on a change in status from their baseline functioning (Aylward, Burt & Thorpe, 1997). For instance, the cognitive decline must interfere with the individual's previous level of social or occupational functioning. Because evidence of decline in previous abilities is necessary, a personal knowledge of the individual is invaluable to the clinician to establish a diagnosis. Unfortunately, because individuals with MR often have poor to no communication skills and loss of speech is a common behavioral symptom of dementia, use of self-report methods is often impossible (Aylward, Burt & Thorpe, 1997). Impairments in verbal skills make it

difficult for many individuals with developmental disabilities to articulate abstract or global concepts such as confusion and disorganized mental abilities. According to Haveman et al.'s (1994) study, approximately 12% of residents in a large-scale residential facility in the Netherlands could not express themselves verbally and showed no signs of comprehension. Another 27% were very restricted in their communication skills.

Self report may be inappropriate for even those individuals with intact expressive language skills because researchers have demonstrated that often, as dementia becomes more severe, patients become less aware of their memory impairment (McDaniel et al., 1995). Therefore, a caregiver's report may be more accurate than information obtained from the individual. Sevush (1999) found that caregivers' evaluation of the patients' memory had better associations with the patients' dementia status and tested cognitive performance than the patients' own evaluation. Therefore, an alternative to self-report is the use of caregiver interviews and informant questionnaires (Zelinski & Gilewski, 1988). Standardized instruments that have been found to be useful for eliciting information from caregivers include the Dementia Questionnaire for Mentally Retarded Persons (DMR; Evenhuis, Kengen, & Eurlings, 1990;

Evenhuis, 1992, 1996), the Dementia Scale for Down's Syndrome (DSDS; Gedye, 1995), and the Early Signs of Dementia Checklist (Visser & Kuilman, 1990).

These interviews should be completed with individuals who are familiar with the person's everyday behavior (Gedye, 1995). The informants should know the person well and have a significant amount of contact with him/her. Someone who was familiar with the person prior to the dementing process is always preferable. This method allows the clinician to establish the symptoms' mode of onset (abrupt versus gradual), progression (stepwise versus continuous decline, worsening versus fluctuating versus improving), and duration. This retrospective information is also critical to determine whether there has been a change in baseline functioning. When the individual lives in an institution or group home with multiple caregivers, it is recommended that multiple informants be interviewed (Gedye, 1998).

Information from individuals familiar with the patient is beneficial but caregiver reports should not be the only source of data. Researchers have found that there are often incongruencies between caregiver reports and objective test measures, with some informants over-reporting the severity of symptoms and others under-

reporting impairment. Prosch-Huy (2001) found significant differences between informant reports and scores on the Folstein Mini Mental State Exam (MMSE), with impairment appearing more severe when measured by MMSE than caregiver report. In contrast, DeBettignies, Mahurin, and Pirozzolo (1993) found that caregivers of adults with Alzheimer's disease rated the patients as being more functionally impaired than what was revealed by actual performance testing.

Because informant reports may be unreliable and inconsistent and retrospective reports may be flawed (especially reports about memory or cognitive functioning), direct assessment of the individual is critical to supplement information supplied by caregivers. The most objective way to measure changes in cognitive and adaptive functioning is the longitudinal administration of tests that assess level of impairment (Aylward, Burt, Thorpe, Lia, & Dalton, 1995). In order to do this; the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disabilities recommended that all adults with mental retardation undergo a comprehensive evaluation (i.e., intellectual assessment, evaluation of adaptive functioning, assessment of psychopathology, and mental status exam) at least once by

the age of 25 years (Aylward et al., 1995). This allows the clinician to establish a record of baseline functioning (Burt & Aylward, 1999).

Cognitive Assessment

Standardized, individually administered intelligence tests are the most important instruments used to diagnose mental retardation (Sattler, 1992). These tests can also be used to track changes in cognitive abilities by comparing the individual's performance over time. Unfortunately, dementing processes are often difficult to identify among persons with intellectual disabilities because subsequent cognitive impairments can be indiscernible (Shultz et al., 1998). Current standardized measures of intellectual functioning may be uninformative, especially in people with very low IQ, as the score may already be so low that no changes are observed with a further dementing process (Janicki, Heller, Seltzer, & Hogg, 1996). Dalton, Seltzer, Adlin, and Wisniewski (1993) attempted to detect cognitive deficits indicative of dementia in persons with Down syndrome. They found that commonly used instruments for assessing cognitive functioning are unreliable in persons with lower cognitive abilities. These measures are often insensitive at the lower end of the cognitive spectrum often causing

individuals with MR to score very poorly due to the so called “floor effect” (Sattler, 1988).

Additionally, many of the mental status exams that are used to evaluate dementia in the general population are not appropriate for use with persons with mental retardation. The mental status exams were designed for individuals whose previous level of cognitive functioning was assumed to be normal (Aylward et al., 1995). Researchers have noted that people who demonstrate “below average” performance on intelligence tests often perform poorly on tests of mental status. These individuals are likely to be labeled “cognitively declined” because of biases built into the measures rather than due to true decline (Shultz, Aman & Rojahn, 1998; Zelinski & Gilewski, 1988). Several mental status examinations have been developed or adapted for use with persons with developmental disabilities. These include the Down’s Syndrome Mental Status Examination (DSMSE) and the Test of Severe Impairment (TSI). The DSMSE (Haxby, 1989) consists of a battery of neuropsychological tests assessing a broad range of skills, including recall of personal information, orientation to season and day of the week, memory, language, visual-spatial function and praxis. The TSI (Albert & Cohen, 1992) also covers a broad range of cognitive functions, including motor performance,

language comprehension, language production, immediate and delayed memory, general knowledge and conceptualization. These tests can be repeated periodically to compare results over time.

Non-Cognitive Assessment

Because cognitive changes are often not readily discernable, clinicians often rely on behavioral symptoms to determine a diagnosis of dementia. In fact, these “non-cognitive” aspects of dementia are often the first signs reported in individuals with MR (Cooper & Prasher, 1998; Evenhuis, 1990; Janicki et al., 1992). Behavioral changes such as irritability, increased wandering, sleep difficulty, urinary incontinence, loss of interest and anhedonia have been related to the development of dementia in persons with MR (Janicki et al., 1995; Prasher, Krishnan, & Clarke, 1994). Other behavioral symptoms that may be indicative of dementia include changes from previously higher levels of adaptive functioning, loss of speech, seizures in previously unaffected individuals, disorientation, and increase in stereotyped behavior (Aitken, Simpson & Burns, 1999; Bozzola, Gorelick, & Freels et al., 1992; Cummings et al., 1994; Janicki et al., 1995). The challenging behaviors displayed by older adults with

dementia may be separated into two categories: behavioral deficits and behavioral excesses (Burgio, 1996).

Behavioral deficits, wherein the non-occurrence of a behavior constitutes a problem, are key defining characteristics of mental retardation. These deficits may have been noted in functional and social domains since childhood (Aylward et al., 1995). However, individuals with mental retardation may present with further decline as they age (e.g., lose ability to perform daily living skills that were previously mastered, become less verbal, and limit social interaction). Declines in social and adaptive functioning beyond baseline levels are a cardinal diagnostic feature of dementia (Shultz, Aman & Rojahn, 1998).

Although some decline in adaptive functioning is expected as an effect of normal aging, decline in the ability to perform activities of daily living is even more pronounced among individuals with dementia. For instance, Zigman et al. (1994) found the level of adaptive functioning significantly decreased in individuals with Down's syndrome aged 50 years and older. Schupf, Lubin and Silverman (1987) analyzed records of 2144 individuals with Down's syndrome and of 4172 controls diagnosed with mental retardation of other etiology. They found that adaptive

competence declined with increasing age to a greater extent for individuals with Down's syndrome than for controls with other intellectual disabilities. The declines noted by Zigman et al. (1994) and Schupf and associates (1987) is presumably attributed to the presence of dementia among aging persons with Down's syndrome. It is known that all individuals with Down's syndrome develop the characteristic neuropathological brain lesions of Alzheimer's disease (i.e., neuritic plaques, granulo-vacuolar changes, cerebral vascular amyloidosis, Hirano bodies and neurofibrillary tangles) by the age of 40 years (Lai & Williams, 1989).

Individuals with Down's syndrome are not the only individuals who present with declines in adaptive behavior. Draper et al. (2000) found that subjects with dementia had greater functional impairments than controls. For instance, individuals with dementia participated in less social activities. Norbergh et al. (2001) investigated the activity of demented patients at a psycho-geriatric unit. The findings showed that persons with dementia who reside in an institution often experience lives of solitude. Armstrong-Esther and Brown (1996) found similar results among patients in a geriatric ward where the individuals spent 88.5% of their time alone. Low social engagement, decreased initiation and lack of interest are common

behavioral symptoms associated with dementia. Apathy, which broadly defines these symptoms, occurs in up to 92% of individuals diagnosed with dementia (Mega et al., 1996).

Researchers have also found that the presence of dementia has dramatic impact on one's communication skills. Draper and colleagues (2000) found that expressive communication skills were significantly more impaired in persons with dementia with only 40% being able to communicate “well enough most times” as compared to 80% of controls. One’s ability to communicate effectively is very important. It influences an individual’s capacity to perform daily living skills, their ability to get their needs met, and has dramatic impact on a person’s social interactions. Researchers have demonstrated that nursing home staff interact more with individuals who can communicate. For instance, Elkman et al. (1991) found that caregivers seldom go to demented patients who have communication problems just to talk to them, and that less time is spent with those individuals during the various care activities.

Although individuals with dementia often present with loss of speech, vocally disruptive behavior, in addition to physical aggression, is one of the most challenging behaviors for nursing home staff to manage (Everitt et al.,

1991; Whall et al., 1992). Disruptive vocalizations include loud requests for attention, chronic screaming, self-talk, negative remarks, and use of obscenities (Vaccaro, 1990). Physically aggressive behaviors include pushing, spitting, grabbing, kicking, hitting, and other dangerous, assaultive behaviors (Cohen-Mansfield, 1989). Rosen et al. (1994) reported that verbal and physical aggression occurred in approximately 80% of nursing home residents diagnosed with dementia. Physical aggression is more common among moderately to severely demented individuals and occurs most often during daily care routines (Hoeffer et al., 1997). These maladaptive behaviors are often referred to as behavioral excesses because the occurrence of these behaviors constitutes the problem (Cohen-Mansfield et al., 1989).

The main factors that seem to contribute to vocally disruptive and physically aggressive behavior are severe impairment in the performance of daily living skills, pain, and communication difficulties (Cohen-Mansfield & Werner, 1997). If we interpret this from a functional standpoint, all behaviors serve a communicative purpose. Individuals with dementia who are no longer able to express themselves with language may use disruptive behavior to reflect an underlying need or discomfort, or a response to

environmental or physical stimuli. Researchers have demonstrated that staff members' responses to these behaviors often inadvertently reward the individual, thereby increasing or maintaining the current rate of aggressive and/or disruptive behavior (Vaccaro, 1990). For instance, staff members may provide the individual with either positive or negative attention (i.e., comforting the person or attempting to stop the behavior through loud verbal reprimands). Parenthetically, even negative attention is rewarding to someone whose schedule of reinforcement is really lean.

Other symptom clusters that are common among persons diagnosed with dementia are "psychosis" (hallucinations, delusions, paranoia), "depression" (sad appearance, crying, guilt, anxiety), and "motor hyperactivity or psychomotor agitation" (pacing, aimless walking, handling objects inappropriately) (Aitkin, Simpson & Burns, 1999). These "non-cognitive" symptoms are commonly associated with the degenerative changes of dementia and they are often the first or only identifiable signs of dementia in persons with developmental disabilities (Moss & Patel, 1995). These behavioral challenges often increase in severity and frequency as the dementia progresses (Cooper et al., 1990; Jacomb et al., 1994).

Rationale and Purpose

Short of autopsy, there are no conclusive biological markers for the most common forms of dementia (Rogan & Lippa, 2002). Therefore, diagnosis is often based on objective longitudinal evidence of deterioration of cognitive abilities and evidence of deterioration in adaptive behavior and/or social skills especially in persons with MR (Aylward et al., 1995). Although these longitudinal changes are essential to diagnose dementia (APA, 1994), groups of investigators have, to some extent, developed their own classification methods (Aylward, Burt & Thorpe, 1997; Barcikowska, Silverman & Zigman, 1989; Burt & Aylward, 1999; Janicki, Heller & Hogg, 1996; Prasher, Krishnan & Clarke, 1994; Shultz, Aman, & Rojahn, 1998; Silverman et al, 1998; Visser & Kuilman, 1990; Visser et al., 1997). Although these approaches share some commonalities (e.g., investigating decline in functioning, comparison between identified “dementia” and “non-dementia” groups) they differ in their emphasis on different types of data (e.g., use of various adaptive behavior measures, mental status examinations, cognitive assessments, scales of psychopathology, and caregiver reports) and analysis (e.g., cross-sectional designs, longitudinal studies).

Burt and Aylward (2000) considered a working battery of tests for the diagnosis of dementia among persons with intellectual disabilities. The battery is divided into two parts: administration of informant-report scales and direct assessment of the individual with MR (Aylward et al., 1997). Administration of six informant report scales is recommended. These include the Dementia Questionnaire for Mentally Retarded Persons, the Dementia Scale for Down Syndrome, the Reiss Screen for Maladaptive Behavior, the Scales of Independent Behavior – Revised, the AAMR Adaptive Behavior Scale – Residential and Community (2nd Ed.), and the Stress Index (Burt & Aylward, 2000). The administration time for each of these scales varies (i.e., according to their respective manuals, the AAMR Adaptive Behavior Scale takes approximately 30 minutes to administer (Nihira, Leland & Lambert, 1993) and the Scales of Independent Behavior – R (Bruinink, Woodcock, Weatherman & Hill, 1996) takes approximately 60 minutes). Administration is manageable when one or two of these scales are used; however, administration of all six scales is quite time-consuming. Burt and Aylward (2000) also selected 10 instruments to be administered directly to the individual with MR (the Test for Severe Impairment, Stanford Binet Sentences, the Fuld (modified),

Autobiographical Memory, Orientation, the Boston Naming Test, the McCarthy Verbal Fluency, Simple Commands, the Purdue Pegboard (modified), and the Developmental Test of Visual Motor Integration). The researchers noted that all of the tests have the capacity to assess individuals with MR ranging from mild to profound levels but this statement can be disputed. For example, several tests recommended are inappropriate for individuals with limited to no verbal skills. According to "administration notes" provided by Burt and Aylward (2000), four of the ten instruments require "speech which is clear enough to score." Also, persons with receptive language deficits may have difficulty comprehending task instructions. Persons with fine motor difficulties are likely to perform poorly on three instruments that require manipulation of small objects and the ability to maneuver a pencil across paper. In addition, it is likely that individuals with short attention spans would have difficulty completing the battery in a single sitting therefore multiple sessions would be required. Fatigue and frustration would also make completion of the battery challenging. Therefore, it is believed that though these instruments may be effective in assessing dementia among individuals with mild and moderate

MR, use with persons with severe and profound MR is dubious.

Visser et al. (1997) also proposed a method to detect dementia in individuals with Down's syndrome. Visser et al.'s (1997) clinical diagnosis of dementia is based on specific clinical symptoms, changes in social skills, and changes in the background alpha rhythm measured by the EEG. The researchers categorized 307 participants into five groups based on the participant's performance on the Early Signs of Dementia Checklist (ESDC, Visser & Kuilman, 1990), scores on the Social Skills Inventory for the Mentally Retarded (Sociale Redzaambeids-shaalvoor Zwakzinnigan; Kraijer & Kema, 1981) and EEG recordings as follows: Group 1 = no deterioration, Group 2 = onset of deterioration, Group 3 = distinct behavioral change, Group 4 = advanced dementia and Group 5 = time of complete dependency. Visser et al. (1997) established the validity of this method by comparing their clinical findings with postmortem neuropathological findings. Permission was obtained to perform an autopsy on 16 patients who died during the course of the study (13 identified with clinical dementia, 3 did not meet Visser et al. (1997) criteria for dementia). A very strong correlation was found. The 13 patients who were clinically diagnosed as having dementia using their

method also presented with brain changes consistent with severe forms of Alzheimer's disease. For instance, senile plaques, neurofibrillary degeneration at the frontal and temporal cortices, severely disrupted interneuronal networks, and neurofibrillary degeneration in the parahippocampal gyrus and the hippocampus were found. Neuroanatomic evidence of Alzheimer disease was not in the autopsy findings in the three other deceased participants who did not meet clinical criteria of dementia.

One particular advantage of the Visser et al. (1997) model is its suitability for use on individuals who have a low level of intellectual functioning. The collection of information about aspects of cognitive functioning is not time consuming and the measures were found to be patient-friendly because they do not involve assessment interaction with the individuals. Another advantage is the converging vectors looking at resident behavior via serial evaluations of participants, caregiver reports with the Early Signs of Dementia Checklist, and the physiological exam findings with the EEG. When all three of these vectors converge, good confidence can be placed in the clinical diagnosis of dementia.

The purpose of this present study was to apply Visser et al.'s (1997) model to determine whether it could detect

dementia among persons diagnosed with severe or profound mental retardation using a social skills measure in common use in facilities in the U.S. The primary reason for focusing on this group of individuals is because persons with severe and profound mental retardation demonstrate the greatest communication deficits and lowest baseline abilities thereby making the identification of dementia based on standard methods such as cognitive assessments and mental status exams quite difficult. This study is more than just a replication of Visser et al.'s research as individuals with and without Down's syndrome were sampled. Much of the research on dementia among persons with MR has been conducted with individuals with Down's syndrome. It is felt that additional studies need to be conducted comparing the prevalence and course of dementia among persons with and without Down's syndrome. In addition, the Matson Evaluation of Social Skills in Individuals with Severe Retardation (MESSIER; Matson, 1995) was used rather than the Social Skills Inventory for the Mentally Retarded. The Social Skills Inventory for the Mentally Retarded does not have an English-version. Several steps would need to be completed before use of this measure would be appropriate in the United States (e.g., translation, cultural adaptation, validation for use in the U.S.,

assessment of the psychometric equivalencies across English and Dutch versions). This process is very time consuming and can be avoided if it can be validly replaced with another well established measure that assesses social skills in individuals with MR. The MESSIER was chosen because it has demonstrated itself to be a reliable and valid measure of social skills in persons with severe and profound MR in the United States.

Visser et al. (1997) were able to gather neuroanatomic evidence of Alzheimer's disease on 13 participants who died during the course of their study. However, postmortem neurological assessments were excluded from this present investigation. Therefore, because of the problem of making an ironclad determination of dementia without evidence of deterioration of brain tissue, a differential prevalence design was used. Differential prevalence designs are often used in research to distinguish malingerers from non-malingerers. This group is another for whom diagnostic confirmations are extraordinarily difficult to obtain. Research on malingering often involves groups of people claiming brain dysfunction who are in litigation with groups of people claiming brain dysfunction who are not in litigation, under the assumption those who are in litigation may be malingering and those who are not in

litigation are not (Greve et al., 2003). Differential prevalence designs are not a guarantee of true assignment. For instance, some individuals grouped as “non-malingering” may indeed be malingering and some grouped as malingerers may be truthful. The problem is that there is no “gold standard” and assignment to “known groups” is only possible if a malingerer is “caught”. Researchers also believe that those caught do not represent the true population of malingerers. Rather, they likely represent unsophisticated malingerers who are easy to detect (Millis, Ross & Ricker, 1998). Using a differential prevalence design for this particular study enables us to compare three groups of individuals who vary in risk for dementia.

Validating Visser et al.’s (1997) method on an American sample is important in light of the dramatic increase of the elderly population of adults with MR in the United States (persons susceptible to dementing processes) and the increased interest in the accurate detection of dementia in persons with developmental disabilities. Early detection is important because there are numerous reports in which the course of dementia can be arrested and in some cases even reversed. Thase (1982) reported a case of reversible dementia secondary to hypothyroidism. A complete psychological/physical assessment indicated

dementia secondary to low thyroid functioning. The patient fully recovered to premorbid functioning when the appropriate treatment was administered. Gedye (1998) described four cases of neuroleptic induced dementia among individuals with intellectual disabilities. These individuals returned to their previous functional states when the offending medications were discontinued.

Early identification of dementia has important clinical implications because it can guide treatment planning and clinical care. Clinicians can use this data to identify decline in previous capabilities and then intervene in order to retrain these skills or prevent further loss. In addition, this data can be used to identify an individual's spared abilities and treatment strategies can be designed to further tap into and take advantage of these facilities. It is believed that good clinical data on the early signs and course of dementia will eventually result in a better outcome for aging persons with mental retardation.

Hypothesis

Hypothesis #1 - It was hypothesized that individuals who belonged to the “high risk” group would demonstrate fewer positive social skills (as evinced by low MESSIER Positive scores and high MESSIER Negative scores) and more characteristic symptoms of dementia (as noted by ESDC item endorsements) than persons in the “medium risk” and “low risk” groups. Individuals identified as “medium risk” would perform worse on the MESSIER Positive and Negative scales and ESDC than persons in the “low risk” group.

Hypothesis #2 – It was hypothesized that individuals with Down’s syndrome would perform worse than persons without Down’s syndrome across the “high” and “medium risk” levels. However, an interaction was predicted, in that Hypothesis #2 would not hold in the “low risk” group. That is, young persons with and without Down’s syndrome would demonstrate little symptoms characteristic of dementia.

Hypothesis #3 – It was hypothesized that the rate of decline in social abilities would be greatest for participants with Down’s syndrome who had been identified as “high risk”.

Method

Participants

Participants were 90 individuals residing at a large developmental center in Louisiana. The sample size was based on results from an a priori power analysis with alpha = .05, power = .80 and the effect size = .30. This effect size is based on an average of several effect sizes reported in dementia research (Burt et al., 1995; Prasher & Chung, 1996; Zigman et al., 2004). All participants were classified, prior to the study, as having severe or profound mental retardation based on DSM-IV criteria (APA, 1994). Fifty-nine of the ninety participants (65.5%) were diagnosed with profound mental retardation.

Using a differential prevalence design, participants were equivalently selected for assignment to one of three groups based on their perceived “risk for dementia”. These groups were selected with the intention that assigning participants to three extreme groups would increase power and likely effect. The high-risk group comprised of 30 individuals who had been referred for psychological assessment in order to screen/rule out for dementia. These individuals had been referred by direct care staff members or by psychology staff because of observed behavioral, cognitive, personality, or social skill changes. No age

limitations were imposed on this group therefore individuals in the high-risk group ranged from 41 years to 92 years, with a mean age of 57.1 years. The medium risk group was made up of 30 “old age” individuals who had not been referred for evaluation. “Old age” was defined as persons aged 55 years or older. Age 55 was selected because researchers have generally defined “old age” in persons with MR as those individuals 55 years and older (Seltzer, 1992). This age cut off seems arbitrary but the criteria are based on several factors including attempts to include some subgroups that seem to age prematurely (i.e., individuals with Down’s Syndrome and individuals with Cerebral Palsy), observations of changes of functions among individuals with MR, and expectations for change in normal age related activities (Seltzer, 1992; Seltzer & Krauss, 1987). These individuals represented the aging population of persons with MR who had not been referred by caretakers due to observed decline/change. The mean age of the medium risk group was 59.9 years, with individuals ranging from 55 to 68 years. The low risk group was comprised of 30 individuals aged 40 years or younger (mean age = 36.1) who had not been referred for a neuropsychological assessment.

One half of the participants for each of the three risk groups were comprised of individuals who have Down’s

syndrome (DS) (n = 45). Every Down's syndrome subject was identified by their characteristic facial features and medical records establishing Down's syndrome.

Table 1
Group Assignment

	High Risk	Medium Risk	Low Risk
Down Syndrome	n=15	n=15	n=15
Non-DS	n=15	n=15	n=15

An attempt was made to match the groups on four factors: gender, level of mental retardation (severe or profound), other psychiatric diagnoses, and psychotropic medication. It is important to match the individuals on "other psychiatric diagnoses" in order to confidently state that significant differences in scores on measures of social skills and current cognitive functioning are a factor of a dementing process rather than the person's co-morbid Axis I diagnosis. Participants were matched on medication to minimize the possibility that loss of skills is due to medication side effects (i.e., lethargy, physical discomfort, etc.).

Interviewers and Informants

All interviewers were master's level psychologists with training specific to the area of mental retardation. Interviewers were trained in the administration of the

assessment instruments by a licensed supervising psychologist or a trained doctoral student in clinical psychology, and had been employed at the developmental center for at least one month prior to conducting interviews. Informants (the individual providing answers to the assessment questions) were direct support staff who had worked with the participants for a minimum of 12 months. All informants held the title of either home manager (charge staff) or group leader.

Table 2
Demographic Variables

		Age	MR Level	Participants with Axis I diagnosis	Participants on psychotropic meds
High Risk	Down's Syndrome	50.7 yrs	Profound = 11 Severe = 4	5	3
	No Down's	63.5 yrs	Profound = 11 Severe = 4	6	6
Medium Risk	Down's Syndrome	58.2 yrs	Profound = 10 Severe = 5	5	2
	No Down's	61.6 yrs	Profound = 9 Severe = 6	5	4
Low Risk	Down's Syndrome	36.5 yrs	Profound = 9 Severe = 6	5	3
	No Down's	35.7 yrs	Profound = 9 Severe = 6	7	4

Predictive Measures

Matson Evaluation of Social Skills in Individuals with Severe Retardation. The Matson Evaluation of Social Skills

for Individuals with Severe Retardation (MESSIER; Matson, 1995) was used as a measure of social skills. The MESSIER was specifically designed to assess social skills in persons with severe and profound mental retardation. It consists of 85 items generated from a review of existing social skills measures for children and adults, items from the social and communication domains of adaptive behavior scales, and items nominated by experts. The items are grouped into six clinically derived subscales: 1) positive verbal, 2) positive non-verbal, 3) positive general, 4) negative verbal, 5) negative non-verbal, and 6) negative general. Each item is rated on frequency using a four-point Likert: never (0), rarely (1), sometimes (2), and often (3). The MESSIER is administered by a trained examiner in a semi-structured interview format. The respondent should be a caregiver who has worked with the individual for at least six months.

The psychometric properties of the MESSIER have been studied. Internal consistence and test-retest reliability was high. Good correlations were also found between raters for the total MESSIER score and for all positive and negative MESSIER items (Matson, LeBlanc, Weinheimer & Cherry, 1999). The convergent validity of the MESSIER was evaluated by comparing it to the Socialization domain of

the VABS and sociometric ratings. Significant positive correlations were found between corresponding MESSIER subscales and VABS subdomains on social behaviors. Although the MESSIER and VABS (Socialization domain) seem to measure similar constructs, the MESSIER was used in this study because it has several advantages over the VABS in measuring social skills in persons with severe and profound mental retardation. First, the MESSIER has a large number of social skills specific questions than the VABS. Second, the MESSIER can be used to illustrate an individual's social skills and deficits, as well as maladaptive behavior excesses, whereas the VABS is limited to providing information about social abilities only.

Early Signs of Dementia Checklist (ESDC; Visser & Kuilman, 1990). The ESDC is a scale designed to assess for clinical signs of mental deterioration. The instrument consists of 37 questions divided over 9 categories (General, Personality Changes, Decrease in Performance, Deterioration of Language Skills, Deterioration of Gait, Disorientation, Incontinence, Epilepsy, and Loss of School-Acquired Skills). These categories were selected to inquire about symptoms that have been found to occur with deterioration associated with dementia (Ballard et al., 2001). There are, however, some troublesome items that may

cause clinicians and researchers to over-predict dementia. For instance, items are endorsed if an individual is currently incontinent. These endorsements may give the impression that deterioration has occurred (i.e., it may appear that an individuals who was able to toilet independently is now having toileting accidents). However, it does not take into account that some persons with limited self-help skills may have never acquired self-toileting skills and may have always relied on protective undergarments and caregiver assistance. Therefore, items on the Incontinence section should be interpreted with caution. It is suggested that items should only be endorsed if there is loss of continence (e.g., a change).

The ESDC is completed via interview. The interviewer should be a trained examiner familiar with the measure. The respondent should be a caregiver who is familiar with the individual and has worked directly with them for at least 6 months. Each item is scored on a binomial scale (Yes- 1, No – 0).

The ESDC has demonstrated good psychometric properties (internal consistency of 0.82 and an interrater reliability of about 0.80). It has also shown to be effective to reliably detect Alzheimer-type dementia at an early stage

when combined with a social skills inventory and EEG (Visser et al., 1997).

Procedure

Retrospective data were utilized to ascertain decline in social skills. This method was accomplished by conducting a chart review of the previous three annual psychological evaluations for each participant and noting the raw scores on the MESSIER Positive and MESSIER Negative scales. Data systems had been set up at the developmental center several years ago with the intention of using the data for research.

There was an approximate one-year interval between administrations of the MESSIER (mean period between administrations of the scale = 10.9 months). This period was selected because, in most instances, a one-year follow-up interval is adequate for monitoring changes in cognitive performance (APA Presidential Task Force, 1998). Visser et al. (1997) noted that all their patients were in a prodromal phase that lasted on average 1.3 years in which the individuals showed aspecific clinical symptoms that heralded the later progressive deterioration. The duration of the deterioration was considered to be the time between the onset of distinctive dementia and complete dependency. The mean duration of deterioration was 2.6 years.

Current testing was completed to collect information about the individual's current functioning via interview with direct care staff using the Early Signs of Dementia Checklist (ESDC). This measure was administered within 3 months of the most recent MESSIER screening in order to limit the possibility of further changes in social skills between test administrations (mean period between administration of MESSIER and ESDC was 2.1 months). The ESDC was used to get a picture of the clinical signs of the individual's deterioration. For this study, the items on the "Incontinence" section were modified to more accurately assess for observed changes in toileting skills. This modification was done to control for persons who may have never acquired independent toileting. The ESDC inquires whether the resident shows urinary incontinence during the day/night either occasionally or continuously and whether the resident shows fecal incontinence. An individual who was never toilet trained may acquire points on the ESDC despite these symptoms being present since baseline and thus, not indicative of a deteriorating process. Therefore, when administering this section, the interviewer was first asked whether the resident was or ever had been able to independently toilet. If the reply was "no", the section was skipped.

Results

Diagnostic Criteria

Following the example of Visser et al. (1997), participants were categorized into one of 5 groups using a cross tabulation procedure. The purpose of this was to show in tabular format the relationship between the 5 categorical values. Those individuals who did not show evidence of decline based on a score of 4 or less on the ESDC were labeled “non-deterioration” (N = 48). A second category “onset of deterioration” was used to describe participants who had a score of 5 or higher on the ESDC but who did not show 25% deterioration in their social skills functioning (N = 27). “Distinct behavioral change” was used to describe individuals who showed 25% deterioration in their positive social skills and a score of 10 or higher on the ESDC (N = 12). “Advanced dementia” was used to categorize persons who showed 50% reduction in positive social skills and at least 10 ESDC symptoms (N = 3). Although some participants had minimal scores on the social skills measure (reflecting the near absence of positive social skills), none of the participants needed nursing care for all domains of daily functioning. Therefore Visser et al.’s (1997) fifth category “complete dependency” was omitted from our analyses.

Next, participants in each of the four Visser et al. categories were sorted further based on their “risk of dementia” status. Of the 48 participants categorized as “no deterioration”, 10 belonged to the high risk group, 15 belonged to the medium risk group, and 23 belonged to the low risk group. Of the 27 participants labeled as “onset of deterioration”, 11 were high risk, 10 were medium risk, and 6 were low risk. Six high risk, 5 medium risk and 1 low risk participant met criteria for “distinct behavior change”. All 3 participants identified as “advanced dementia” belonged to the high risk group.

Table 3
Classification into 5 “Dementia Categories”

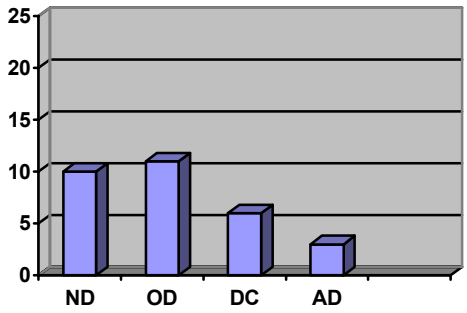
	Non-Deterioration	Onset of Deterioration	Distinct Change	Advanced Dementia
High Risk	10	11	6	3
Medium Risk	15	10	5	0
Low Risk	23	6	1	0

These data demonstrate that, as compared to the medium and low risk groups, individuals classified as high risk had the least number of participants meeting Visser et al. (1997) criteria for “no deterioration” but the greatest prevalence of individuals meeting criteria for “advanced dementia”. Conversely, the low risk group had the greatest

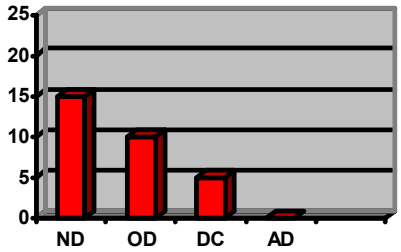
number of participants in the “no deterioration” category and the least in the “distinct change” category.

Legend
ND = No Deterioration
OD = Onset of Deterioration
DC = Distinct Change
AD = Advanced Dementia

HIGH RISK GROUP



MEDIUM RISK GROUP



LOW RISK GROUP

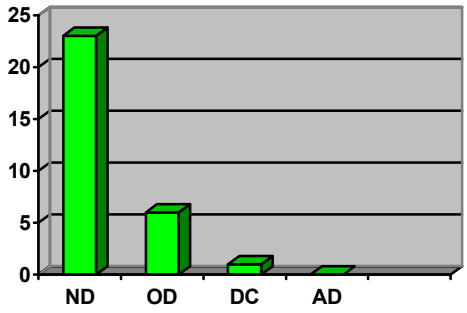


Figure 1
Distribution of Dementia Category Frequencies

Though the cross tabulation graphs look different for the three risk referral groups, a hypothesis test was performed on the contingency table to determine whether or not dementia classification was distributed similarly across the different “risk of dementia” groups. The observed frequencies were used to compute the expected frequencies for the four Visser et al. (1997) categories. It was determined that the expected count of individuals in each of the Visser et al. categories for the high, medium and low risk groups was as follows: “no deterioration” = 16, “onset of deterioration” = 9, “distinct change” = 4, and “advanced dementia” = 1. A chi square test compared the expected and the observed frequencies in each cell. Chi square is most frequently used to test the statistical significance of results reported in a contingency table (Brymer & Cramer, 2001)). The computed chi square statistic (χ) of .012 was less than .05 indicating that assignment to the Visser et al. categories was not distributed similarly across the different levels of risk. The frequency of individuals meeting Visser et al. criteria for dementia classification was related to their perceived risk (e.g., individuals identified as high risk are more likely to meet Visser et al. criteria for dementia than individuals identified as low risk).

Cross Sectional Analyses

A cross sectional analysis (multivariate analysis of variance) was used to examine, at one point in time, differences in ESDC total scores, MESSIER Positive total scores, and MESSIER Negative total scores between persons with and without Down's Syndrome (independent variable #1 – "syndrome") and persons assigned to the three perceived risk groups (independent variable #2 – "risk of dementia"). This analysis yielded significant main effects for "syndrome" ($F(1,88)=4.566$, $p=.005$)and for "risk of dementia" ($F(2,87)=5.844$, $p=.000$). An interaction effect (syndrome * risk of dementia) was not identified ($F(5,84)=1.020$, $p=.414$). Please refer to Table 4 for means and standard deviations.

Tests of between subjects effects indicated that participants with Down's Syndrome ($M=14.22$) had significantly lower scores on the MESSIER Negative scale ($F(1,88)=13.491$, $p=.000$) than participants without Down's Syndrome ($M=25.27$). Significant differences were not noted between the two syndrome groups on either the MESSIER Positive ($F(1,88)=.084$, $p=.772$) or ESDC Total scores ($F(1,88)=1.491$, $p=.225$).

Significant differences on MESSIER Negative ($F(2,87)=3.162$, $p=.047$) and ESDC Total scores

(F(2,87)=18.759, p=.000) were noted among individuals assigned to the three “risk of dementia” groups. A post hoc test indicated significant differences (p=.048) between the high and low risk groups on the MESSIER Negative scale (M=24.567 and M =15.333, respectively).

Analysis revealed that the high risk group (M=8.467) was also significantly different from both the medium risk (M=4.033, p=.001) and low risk group (M=1.600, p=.000) on the ESDC Total score. However, significant differences were not noted between the medium and low risk groups on any of the three assessment scales (MESSIER Positive p=.456, MESSIER Negative p=.557, ESDC p=.107). This indicates that, despite a mean age difference of 23.8 years, scale scores were statistically similar among individuals who were not referred for neuropsychological assessment.

A separate analysis was completed on the total study sample (N=90) in order to investigate possible age effects. Participants were divided into 5 groups according to their chronological ages. Group 1 was comprised of individuals between the ages of 30 and 39 years (N=28, Note: one subject was less than 30 years but they were included in group 1). Group 2 was made up of 12 individuals who were between 40 and 49 years. There were 26 participants

between the ages of 50 and 59 years (group 3) and 19 participants between the ages of 60 and 69 years (group 4). The fifth group contained all individuals who were 70 years and older (n=5). A multivariate analysis of variance was conducted to determine whether there were significant differences across age groups in terms of ESDC, MESSIER Positive, and MESSIER Negative total scores. Results showed that age had a significant effect on the ESDC scores ($F(4,85)=4.651$, $p=.002$). Multiple comparisons were completed in order to determine among which age groups these differences were found. Results showed that the 30-39 year old group differed significantly from the 50-59 year old group ($p=.046$) and from the 60-69 year old group ($p=.031$). Age did not have a significant effect on either the MESSIER Positive ($F(4,85)=.248$, $p=.910$) or MESSIER Negative ($F(4,85)=1.961$, $p=.108$) scores.

Another series of multiple analyses of variance were run to investigate the effects of our participants' demographic characteristics on the scale scores. Two separate analyses were conducted to determine whether there were significant differences across gender groups and levels of mental retardation. A significant difference between males and females was not found on any of the three dependent measures (ESDC – $F(1,88)=.457$, $p=.501$; MESSIER

Positive – F(1,88)=.004, p=.952; MESSIER Negative – F(1,88)=.466, p=.496). However, a significant difference between levels of MR was obtained when examining MESSIER Positive scores (F(1,88)=13.16, p=.000). Individuals with severe MR had significantly higher MESSIER Positive scores (M=102.48, SD=35.27) than participants diagnosed with profound MR (M=76.02, SD=31.59). A significant difference between participants with severe and profound MR was not found on either MESSIER Negative (F(1,88)=.052, p=.821) or ESDC total scores (F(1,88)=.009, p=.923).

Table 4
Means and Standard Deviations for ESDC Total and MESSIER Positive and Negative Scores

Referral Status	Syndrome	MESSIER Positive		MESSIER Negative		ESDC Total	
		Mean	S.D.	Mean	S.D.	Mean	S.D.
High	D.S.	80.267	35.526	17.267	14.655	8.133	7.763
	Non-D.S.	77.067	30.056	31.867	21.112	8.800	5.414
Medium	D.S.	87.267	37.107	14.000	8.036	2.533	2.642
	Non-D.S.	100.800	29.898	24.667	13.500	5.533	3.159
Low	D.S.	91.067	40.013	11.400	11.544	1.733	2.219
	Non-D.S.	74.333	35.407	19.267	13.419	1.467	2.200

Cross Sectional Analysis of Clinical Symptoms

The clinical signs of dementia (as measured by the ESDC) were investigated more closely with frequency counts and a multiple analysis of variance. Results showed that 49% of our participants had at least one item endorsed on the “Decreased Performance” scale of the ESDC. Personality changes were endorsed for 44.4% of participants, indicating recent changes in mood and an exaggeration of personality traits. Approximately one-fourth (27.7%)of our participants presented with aspecific clinical symptoms such as a decrease in interest, speed and motivation. Twenty seven percent of participants were noted to experience some deterioration in gait and 27.8% were beginning to occasionally show urinary incontinence. To a lesser degree, participants showed signs of spatial and temporal disorientation (13.3%) as well as deterioration in language skills (15.6%). Only 6 of the 90 participants demonstrated a loss of school-acquired skills, specifically deterioration in the ability to read and write, and only one participant developed epilepsy during the course of the study.

A multivariate analysis of variance (MANOVA) was conducted in order to establish whether an individual’s “syndrome” or “risk of dementia” status influenced the

items endorsed on the ESDC. The results indicated that a significant difference on the Personality Changes scale existed among participants with DS (M = .8889) and persons without DS (M = 1.4889) ($F(1,88)= 6.231$, $p = .015$).

Significant differences between "syndrome" groups were not noted on any other ESDC scales.

A significant main effect for "risk of dementia" ($F(2,87)=3.382$, $p=.000$) was also noted. Tests of between-subjects effects indicated that significant values were obtained for all ESDC scales, with the exception of the Epilepsy scale ($F(2,87)=1.000$, $p=.372$). Multiple comparisons were made (Sheffe post-hoc test) to identify the specific differences within the groups. Significant differences were noted between the high and low risk groups on the following scales: General ($p=.003$), Personality Changes ($p=.000$), Decreased Performance ($p=.000$), Deterioration of Language Skills ($p=.000$), Deterioration of Gait ($p=.002$), Disorientation ($p=.024$), Incontinence ($p=.025$) and Loss of School Acquired Skills ($p=.030$). Significant differences between the high and medium risk groups were noted on the Decreased Performance ($p=.001$), Deterioration of Language Skills ($p=.000$) and Deterioration of Gait ($p=.006$) scales. A significant difference was

noted between the medium and low risk group on the Personality Changes scale (p=.001).

Longitudinal Analyses

Because reliable diagnosis of dementia requires systematic documentation of age related performance declines within affected people (APA, 1994), a series of repeated measures analyses of variance were used to test for significant changes in social abilities over time. The effects of referral status upon MESSIER Positive and MESSIER Negative scores were investigated with two separate two-way ANOVAs comparing the three “risk of dementia” groups over the three test administrations. Main effects for “time” were noted for the MESSIER Positive ($F(2,87)=4.217$, $p=.016$) and MESSIER Negative scores ($F(2,87)= 3.200$, $p= .043$).However, a significant main effect for “risk of dementia” was not noted for either the MESSIER Positive ($F(2,87)=1.093$, $p=.340$) or MESSIER Negative scores ($F(2,87)= .393$, $p= .676$). Significant interaction effects (Time * Risk of Dementia) were not noted for MESSIER Positive ($F(5,84)=2.003$, $p=.096$) or MESSIER Negative scores ($F(5,84)= 2.001$, $p= .096$).

The effects of syndrome group upon MESSIER Positive and MESSIER Negative scores were also investigated. Main effects for “time” were noted for the MESSIER Positive

(F(1,88)=4.078, p=.019) and MESSIER Negative scores (F(1,88)= 3.105, p= .047). A significant main effect for "syndrome" was noted for MESSIER Negative scores (F(1,88)= 237.147, p= .000) but not for MESSIER Positive scores (F(1,88)=1.093, p=.340). Significant interaction effects (Time * Syndrome) were not noted for MESSIER Positive (F(4,85)=2.003, p=.096) or MESSIER Negative scores (F(4,85)= .288, p= .750).

Table 5
Longitudinal Analysis of MESSIER Positive Scores

		Administration 1	Administration 2	Administration 3
High Risk	DS	M=92.86, SD=29.09	M=87.73, SD=27.18	M=80.27, SD=35.53
	Non-DS	M=91.13, SD=29.65	M=88.60, SD=35.31	M=77.07, SD=30.06
Medium Risk	DS	M=89.20, SD=39.13	M=88.27, SD=35.28	M=87.27, SD=37.11
	Non-DS	M=104.60, SD=35.63	M=98.40, SD=32.90	M=100.80, SD=29.90
Low Risk	DS	M=94.33, SD=40.25	M=89.13, SD=36.39	M=91.07, SD=40.01
	Non-DS	M=74.87, SD=31.93	M=72.60, SD=33.72	M=74.33, SD=35.09

Table 6
Longitudinal Analysis of MESSIER Negative Scores

		Administration 1	Administration 2	Administration 3
High Risk	DS	M=10.93, SD=10.74	M=15.27, SD=16.30	M=17.27, SD=14.66
	Non-DS	M=20.67, SD=14.07	M=22.80, SD=18.70	M=31.87, SD=21.11
Medium Risk	DS	M=11.93, SD=5.84	M=11.67, SD=8.04	M=14.00, SD=8.04
	Non-DS	M=19.07, SD=11.28	M=24.00, SD=14.80	M=24.67, SD=13.50
Low Risk	DS	M=11.53, SD8.04	M=14.47, SD=9.65	M=11.40, SD=11.54
	Non-DS	M=20.40, SD=23.17	M=26.13, SD=24.78	M=19.27, SD=13.42

Discussion

The life expectancy of adults with mental retardation has significantly increased in recent years (Silverman et al., 1998; Yang, Rasmussen & Friedman, 2000). As a result, the prevalence of age-associated diseases such as dementia of the Alzheimer's type has also risen (Zigman, Silverman, & Wisniewski, 1996). Unfortunately, accurate prevalence rates are difficult to obtain because the evaluation and diagnosis of dementia in persons with MR is a complicated and involved process. Due to these individuals' life long histories of cognitive impairment and the potentially unique presentation of clinical symptoms of dementia in this population, valid assessment of dementia in persons with MR remains a problematic challenge. In addition, although an increasing number of researchers are focusing on the aging process in persons with intellectual disabilities, comparison of experimental results is difficult due to the lack of uniform research methods and clear diagnostic criteria. This study set out to replicate the diagnostic method proposed by Visser et al. (1997) on a sample of 90 developmentally disabled individuals with and without Down's syndrome. Prevalence rates and presenting symptoms were compared with Visser et al. (1997) results. Further analysis investigated the effects of age, sex,

level of MR, syndrome, and perceived risk of dementia on the participants' assessment scores.

We discovered that, during the course of three years, 15 (16.7%) of our 90 participants developed symptoms of dementia compatible with Visser et al.'s (1997) criteria. Seven of those participants who met criteria had a co-morbid diagnosis of Down's syndrome. These rates are very similar to results obtained by Visser et al. (1997) that showed that 18% of their patients with Down's syndrome developed symptoms of dementia. Also consistent with the results of Visser et al. (1997), our participants showed greatest decline in their ability to perform coordinated movements, as demonstrated by a poorer work performance, greater dependence on assistance to complete daily living skills, and onset of deterioration in their ability to perform household chores. We acknowledge that, without postmortem examination, we cannot guarantee that our 15 participants were truly showing symptoms indicative of dementia. Visser et al. (1997) was able to provide more diagnostic confirmation by including EEG recordings in their screening. Further confirmation was obtained for Visser at al. (1997) when a marked association was noted between their clinical diagnosis and neurological changes in the brains of 13 patients who died during the course of

their study. However, because our methodology and diagnostic criteria were similar to Visser et al.'s (1997) study, we feel that we have successfully replicated their results and have demonstrated that the MESSIER is an appropriate substitute for the Social Skills Inventory for the Mentally Retarded.

Using a differential prevalence design we were able to compare our three risk groups (high = those referred by caretakers, medium = individuals 55 years and older who had not been referred, and low = individuals 40 years and younger who had not been referred). Results indicated that the degree of deterioration of skills was significantly different among the groups, with the greatest proportion of persons demonstrating symptoms of dementia belonging to the high risk group. However, it should be noted that though the Pearson chi-square statistic was significant (p=.012), it is possible that a difference was predicted that was not actually there. Statisticians recommend that the chi square test not be used if any cell has an expected frequency of less than one or if more than 20% of the cells have an expected frequency of less than 5. Though our expected frequencies were not less than one, six of our 12 cells (50%) had an expected count less than 5. Thereby our cell frequencies were too low for chi square to be

appropriately used. We could have placed greater confidence on our results if our sample size had been larger, thereby increasing the number of individuals in each cell. However, because the number of individuals with Down's syndrome that resided in the developmental center limited our total sample size to 90, it is suggested that data continue to be collected on the present sample. The rationale for this is to determine whether our participants' skills continue to deteriorate and whether the prevalence of individuals showing symptoms of dementia increases. The length of our study was less than a third of Visser et al.'s (1997) 10-year study. Three years may not be sufficient to observe significant declines. This will be discussed further in reference to the results of our longitudinal analysis.

In addition to making a comparison with Visser et al.'s (1997) study, this study was designed to examine age-related changes in behavioral and social functioning in individuals with and without Down's syndrome over a three year period. A number of recently published studies have established an association between Alzheimer's dementia and Down's syndrome (Crayton, Oliver, Holland, Bradbury & Hall, 1998; Devenny, Krinsky-McHale, Serson, & Silverman, 2000; Holland, Hon, Huppert & Stevens, 2000). Therefore, it was

hypothesized that persons with Down's syndrome, particularly those who had been referred for a neurological assessment, would have higher scores on the ESDC total and MESSIER Negative scale, as well as lower MESSIER Positive scores than participants that did not have a co-morbid diagnosis of Down's syndrome. However, our results were inconsistent with studies that have found an accelerated rate of decline for persons with Down's syndrome as compared to other individuals with mental retardation (Das, Mishra, Davison & Naglieri, 1995; Thompson, 2003). Significant differences between the syndrome groups were not noted in either ESDC total or MESSIER Positive scores. There are two possible explanations why differences between our "syndrome" groups were not observed. Both explanations focus on our subjects' level of mental retardation and how pre-morbid cognitive deficits can effect the scores of persons with and persons without Down's syndrome.

One explanation for the absence of significant differences between the "syndrome" groups is the effect of pre-morbid deficits upon the prevalence rates of our participants with Down's syndrome. In this present study, 15.5% of our Down's syndrome sample showed symptoms of dementia. This is significantly lower than prevalence rates noted in the literature (Zigman, Schupf, Haveman &

Silverman, 1996). For instance Holland et al. (2000) found that approximately 60% of their participants with Down's syndrome developed dementia by the time they reached 60 years of age. Unlike other studies that included persons functioning within the mild and moderate ranges of MR (Das, Mishra, Davison & Naglieri, 1995), this present study only contained persons with severe and profound MR. It is believed that declines were not as evident for our sample because their low pre-morbid abilities made identification of further decline very difficult. Deterioration in skills may have appeared small and slow because there was not enough room for further decline.

The second, and more likely explanation considers the effect of pre-morbid functioning upon the non-Down's syndrome group. Although significantly fewer studies have looked at dementia in persons without Down's syndrome, researchers have established much lower rates for this population in comparison to prevalence rates of persons with Down's syndrome. For instance, Janicki and Dalton (2000) found that 6.1% of their adults with MR but without Down's syndrome were classified with dementia. However, more than 17% of our participants demonstrated symptoms of dementia. This accelerated rate may be explained by the "Reserve Capacity Model" (Mortimer, 1988). The reserve

capacity model is based on research that shows that brain cells gradually die as people age (Jorm, 1996). However, because most individuals have a sufficient "reserve capacity" of brain cells to compensate for these losses, declines in cognitive functioning are not immediately evident. Signs and symptoms of dementia only appear when an excess of neurons are lost and compensation is no longer possible (Katzman, 1993). The reserve capacity model states that persons who already have low levels of cognitive abilities (and for whom initial cognitive capacity is likely to be diminished) are at increased risk for dementia (Snowdon, Greiner, Kemper, Nanayakkara & Mortimer, 1999; Whalley et al., 2000). Furthermore, individuals with lower levels of functioning (our subjects with severe and profound MR) are expected to experience an earlier onset of dementia symptoms and a faster rate of decline (Devenny et al., 1996; Temple, Joszvaki, Konstantareas & Hewitt, 2001). It is hypothesized that individuals with severe and profound MR have accelerated rates of dementia because of limited brain cell reserve. The additional diagnosis of Down's syndrome has little effect on persons already functioning at these low pre-morbid states.

The only difference that was noted between our "syndrome" groups was on the MESSIER Negative scale. We had predicted that, consistent with previous studies (Thompson, 1999), our Down's syndrome group would show a greater amount of negative social behaviors than our non-Down's syndrome group. However, the converse was actually found and our non-Down's syndrome group demonstrated more negative behaviors. We feel this may be related to behavioral phenotypes that have been associated with Down's syndrome (Dykens & Kasari, 1997; Kasari & Freeman, 2001). Studies have shown that individuals with Down's syndrome tend to be more social than individuals with MR due to other etiologies. For instance, Kasari and Hodapp (1996) found that children with Down's syndrome displayed greater amounts of social, engaging behavior than were typically found in other types of retardation. These behavioral phenotypes may persist throughout the individual's lifespan and make development of negative social behaviors less likely.

These cross sectional analyses are, at best, an indirect reflection of the differences in these samples. Therefore caution needs to be used when interpreting these data because of methodological limitations associated with cross sectional studies (e.g., cohort effects and selective

survival). In fact, it is believed that selective survival may have prevented us from observing the age effects that are common in dementia studies. Selective survival is based on the notion that older age groups consist of a pool of surviving subjects and these survivors may have been individuals whose average health at age 40 was better than those who did not survive to old age (Strauss & Zigman, 1996; Widaman et al, 1994). This “selective survival” effect is likely to be a problem among adults with MR who are known to experience premature mortality (Burt et al., 1995). Our comparison of the medium (“old age”) and low risk group did not result in any significant differences in ESDC, MESSIER Positive or MESSIER Negative scores. It is believed that, due to selective survival, our sample of older adults (medium risk group) may have been more fit than our sample of younger adults (low risk group). Thus, our older group may have been unrepresentatively healthy whereas it is likely that our younger group was made up of future survivors and non-survivors. Further evidence of selective survival is demonstrated by the scores of our Down’s syndrome participant who was over the age of 70. This individual had no items endorsed on the ESDC, his MESSIER Positive score was higher than the mean of any

other age group, and his MESSIER Negative score was lower than the mean of any other age group.

Researchers have long established that the preferred design for a study of aging is a longitudinal design. In such a design, cohort effects would not be confounded with age effects (Salthouse, 1982). A repeated measure ANOVA was used to test for significant changes in MESSIER Positive and MESSIER Negative scores over time with the expectation that longitudinal changes would not be evident in the low risk group, would be evident in the medium risk group, and would be most evident in the high risk group. Changes in scores were noted over the three test administrations but the rate of decline was statistically similar across all groups regardless of their “risk of dementia” or “syndrome” status.

The length of our longitudinal study may provide a possible explanation why significant main effects for “syndrome” and “risk of dementia” or significant interaction effects for “time * syndrome” and “time * risk of dementia” were not found. Previous studies that have shown differential rates were conducted for a longer duration (e.g., Visser et al. (1997) monitored their participants for up to 10 years). Or, the period between pre- and post-tests was longer (e.g., Holland and Hon

(1996) waited 18 months before re-administering their assessments). Once again, it is suggested that data continue to be collected on this sample to determine whether the rates of decline continue to change as time passes.

This present study had several limitations. As mentioned, the duration of the study may not have been long enough and one year between test administrations may not have been sufficient enough to see a decline in skills. A second limitation was our sample size. We had difficulty finding potential participants with Down's syndrome who fit our criteria (persons with severe or profound MR, individuals between the ages of 25-40 or 55 years and older) so this ultimately resulted in small cell sizes in our bivariate table. Also, because this study only involved an institutionalized population, our results cannot be directly projected to non-institutionalized individuals with mental retardation. The greatest limitation is that we have no pathology findings for any of our cases that show symptoms of dementia. Therefore, although we can hypothesize that the observed declines in social and behavioral functioning are due to dementia, we cannot confirm this without autopsy findings.

Overall, results of this study indicate that the Visser et al. (1997) method was useful in distinguishing between “risk of dementia” groups. Although differences between syndrome groups were not as large as anticipated, valuable information was obtained regarding the signs, symptoms and course of dementia in persons with intellectual disabilities. In addition, this study provides further support that, though caregiver reports are useful in identifying functional impairments, clinicians should not wait for a referral before completing a dementia assessment on an aging individual. Though the high risk group (those referred by caregivers) contained the largest proportion of persons meeting Visser et al. (1997) criteria for dementia, testing demonstrated that 10% of non-referred individuals also showed signs of deterioration. Therefore, all individuals with developmental delays should undergo a comprehensive evaluation by the age of 25 years and testing should be repeated at least once every five years to assess for declines in functioning (Aylward et al., 1995).

This is particularly important because assessment may allow clinicians to detect dementia early enough stage to to attempt to slow the individual’s cognitive decline with medication. Although there is no cure for dementia of the Alzheimer’s type, there are a number of drugs available

that have shown effective in stabilizing the symptoms of dementia, thus improving well being and easing caregiver burden. The drugs that have been most widely used include the cholinesterase inhibitors: donepezil (Aricept), galantamine (Reminyl), and rivastigmine (Exelon) and a glutamate blocker: memantine (Namenda). (Reisberg et al., 2003; Winblad & Portis, 1999).

Identification of the onset of deterioration should also influence treatment planning. Training priorities should change once progressive delays are identified. For instance, the focus of treatment can be teaching these individuals alternate modes of communicating their wants and needs and preserving whatever intact communication skills there are. Social skills training may also be beneficial but with a focus of training shifted from new skills acquisition to current skills preservation. This may enhance the quality of their lives by sustaining their ability to participate in interpersonal interactions and thus thwart or even just postpone the course and pace of deterioration (Walsh et al., 2001). In addition, educating staff members of the dementing process and providing guidelines for caring for persons clinically diagnosed with dementia may improve the quality of the clients' lives and enhance the staffs' caregiver roles by allowing their

attitude to change their caregiving activities as clients' needs and abilities change (Engelman, Altus & Matthews, 1999). Thus, changing training priorities can have a dramatic impact on the resident's activities of daily living and perhaps postpone their transfers to a more restrictive mental health setting.

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Vita

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